Agency for Healthcare Research and Quality
Advancing Excellence in Health Care

NATIONAL GUIDELINE CLEARINGHOUSE

General

Guideline Title

Hemoglobinopathies in pregnancy.

Bibliographic Source(s)

American College of Obstetricians and Gynecologists (ACOG). Hemoglobinopathies in pregnancy. Washington (DC): American College of Obstetricians and Gynecologists (ACOG); 2007 Jan. 9 p. (ACOG practice bulletin; no. 78). [26 references]

Guideline Status

This is the current release of the guideline.

The American College of Obstetricians and Gynecologists (ACOG) reaffirmed the currency of this guideline in 2011.

Regulatory Alert

FDA Warning/Regulatory Alert

Note from the National Guideline Clearinghouse: This guideline references a drug(s) for which important revised regulatory and/or warning information has been released.

- March 22, 2016 – Opioid pain medicines : The U.S. Food and Drug Administration (FDA) is warning about several safety issues with the entire class of opioid pain medicines. These safety risks are potentially harmful interactions with numerous other medications, problems with the adrenal glands, and decreased sex hormone levels. They are requiring changes to the labels of all opioid drugs to warn about these risks.

Recommendations

Major Recommendations

The grades of evidence (I-III) and levels of recommendation (A-C) are defined at the end of the "Major Recommendations" field.

The following recommendations are based on good and consistent scientific evidence (Level A):

> Individuals of African, Southeast Asian, and Mediterranean descent are at increased risk for being carriers of hemoglobinopathies and should be offered carrier screening and, if both parents are determined to be carriers, genetic counseling.

A complete blood count and hemoglobin electrophoresis are appropriate laboratory tests for screening for hemoglobinopathies. Solubility tests alone are inadequate for screening because they fail to identify important transmissible hemoglobin gene abnormalities affecting fetal outcome.

Couples at risk for having a child with sickle cell disease or thalassemia should be offered genetic counseling to review prenatal testing and reproduction options. Prenatal diagnosis of hemoglobinopathies is best accomplished by DNA analysis of cultured amniocytes or chorionic villi.

Definitions:

Grades of Evidence

I Evidence obtained from at least one properly designed randomized controlled trial.

II-1 Evidence obtained from well-designed controlled trials without randomization.

II-2 Evidence obtained from well-designed cohort or case–control analytic studies, preferably from more than one center or research group.

II-3 Evidence obtained from multiple time series with or without the intervention. Dramatic results in uncontrolled experiments also could be regarded as this type of evidence.

III Opinions of respected authorities, based on clinical experience, descriptive studies, or reports of expert committees.

Levels of Recommendation

Level A — Recommendations are based on good and consistent scientific evidence.

Level B — Recommendations are based on limited or inconsistent scientific evidence.

Level C — Recommendations are based primarily on consensus and expert opinion.

Clinical Algorithm(s)

The original guideline document contains a clinical algorithm for "Specialized antepartum evaluation for hematologic assessment of patients of African, Southeast Asian, or Mediterranean descent."

Scope

Disease/Condition(s)

Hemoglobinopathies in pregnancy, including:

Sickle cell disease
Thalassemia (alpha and beta)

Guideline Category

Counseling

Management

Risk Assessment

Screening

Clinical Specialty

Hematology

Medical Genetics

Obstetrics and Gynecology

Pediatrics

Intended Users

Physicians

Guideline Objective(s)

To aid practitioners in making decisions about appropriate obstetric and gynecologic care
To provide recommendations for screening and clinical management of hemoglobinopathies during pregnancy

Target Population

Pregnant women with hemoglobinopathy, including sickle cell disease and thalassemia
Couples at risk for having a child with hemoglobinopathies, especially individuals of African, Southeast Asian, and Mediterranean descent

Interventions and Practices Considered

Screening/Risk Assessment

Risk assessment (e.g., ethnicity)
Laboratory testing (complete blood count, hemoglobin levels, hemoglobin electrophoresis, solubility testing for hemoglobin S, isoelectric focusing, high-performance liquid chromatography, mean corpuscular volume, serum ferritin levels)
Genetic testing (parents, fetus via chorionic villus sampling or amniocentesis, preimplantation)

Management

Genetic counseling
Folic acid supplementation
Cesarean delivery (not recommended routinely)
Epidural analgesia
Avoidance of triggers of painful crisis
Analgesia for painful crisis (e.g., opiates)
Detection and treatment of acute chest syndrome, infection, dehydration, severe anemia, cholecystitis, and hypersplenism
Blood transfusion, including prophylactic exchange transfusion
Multidisciplinary management
Antenatal fetal surveillance (serial ultrasound, nonstress test, contraction stress test)

Major Outcomes Considered

Completion of pregnancy
Maternal and fetal perinatal morbidity and mortality

Methodology

Methods Used to Collect/Select the Evidence

Methods Used to Collect/Select the Evidence

Hand-searches of Published Literature (Primary Sources)

Hand-searches of Published Literature (Secondary Sources)

Searches of Electronic Databases

Description of Methods Used to Collect/Select the Evidence

2007 Guideline

The MEDLINE database, the Cochrane Library, and the American College of Obstetricians and Gynecologists' own internal resources and documents were used to conduct a literature search to locate relevant articles published between January 1985 and March 2005. The search was restricted to articles published in the English language. Priority was given to articles reporting results of original research, although review articles and commentaries also were consulted. Abstracts of research presented at symposia and scientific conferences were not considered adequate for inclusion in this document. Guidelines published by organizations or institutions such as the National Institutes of Health and the American College of Obstetricians and Gynecologists were reviewed, and additional studies were located by reviewing bibliographies of identified articles.

2011 Reaffirmation

Medline/Pubmed/Cochrane databases were searched for literature published from 2007-2011.

Number of Source Documents

Not stated

Methods Used to Assess the Quality and Strength of the Evidence

Weighting According to a Rating Scheme (Scheme Given)

Rating Scheme for the Strength of the Evidence

Studies were reviewed and evaluated for quality according to the method outlined by the U.S. Preventive Services Task Force:

I Evidence obtained from at least one properly designed randomized controlled trial.

II-1 Evidence obtained from well-designed controlled trials without randomization.

II-2 Evidence obtained from well-designed cohort or case–control analytic studies, preferably from more than one center or research group.

II-3 Evidence obtained from multiple time series with or without the intervention. Dramatic results in uncontrolled experiments also could be regarded as this type of evidence.

III Opinions of respected authorities, based on clinical experience, descriptive studies, or reports of expert committees.

Methods Used to Analyze the Evidence

Systematic Review

Description of the Methods Used to Analyze the Evidence

Not stated

Methods Used to Formulate the Recommendations

Expert Consensus

Description of Methods Used to Formulate the Recommendations

2007 Guideline

Analysis of available evidence was given priority in formulating recommendations. When reliable research was not available, expert opinions from obstetrician–gynecologists were used. See also the "Rating Scheme for the Strength of Recommendations" field regarding Grade C recommendations.

2011 Reaffirmation

A committee member reviewed the document and new literature search on the topic. The document was then reviewed by the committee and the committee agreed that it is current and accurate.

Rating Scheme for the Strength of the Recommendations

Based on the highest level of evidence found in the data, recommendations are provided and graded according to the following categories:

Level A — Recommendations are based on good and consistent scientific evidence.

Level B — Recommendations are based on limited or inconsistent scientific evidence.

Level C — Recommendations are based primarily on consensus and expert opinion.

Cost Analysis

A formal cost analysis was not performed and published cost analyses were not reviewed.

Method of Guideline Validation

Internal Peer Review

Description of Method of Guideline Validation

Practice Bulletins are validated by two internal clinical review panels composed of practicing obstetrician-gynecologists generalists and sub-specialists. The final guidelines are also reviewed and approved by the American College of Obstetricians and Gynecologists (ACOG) Executive Board.

Evidence Supporting the Recommendations

Type of Evidence Supporting the Recommendations

The type of supporting evidence is identified and graded for each recommendation (see "Major Recommendations").

Benefits/Harms of Implementing the Guideline Recommendations

Potential Benefits

Appropriate screening and management of hemoglobinopathies during pregnancy

Potential Harms

Epidural analgesia usually is well tolerated as long as care is taken to avoid hypotension and hypoxemia.

Contraindications

Contraindications

Hydroxyurea has been shown to reduce the frequency of painful crises in nonpregnant patients with severe sickle cell disease. However, the use of hydroxyurea is not recommended during pregnancy because it is teratogenic.

Qualifying Statements

Qualifying Statements

These guidelines should not be construed as dictating an exclusive course of treatment or procedure. Variations in practice may be warranted based on the needs of the individual patient, resources, and limitations unique to the institution or type of practice.

Implementation of the Guideline

Description of Implementation Strategy

An implementation strategy was not provided.

Implementation Tools

Clinical Algorithm

For information about availability, see the *Availability of Companion Documents* and *Patient Resources* fields below.

Institute of Medicine (IOM) National Healthcare Quality Report Categories

IOM Care Need

Living with Illness

Staying Healthy

IOM Domain

Effectiveness

Patient-centeredness

Identifying Information and Availability

Bibliographic Source(s)

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Adaptation

Not applicable: The guideline was not adapted from another source.

Date Released

2007 Jan (reaffirmed 2011)

Guideline Developer(s)

American College of Obstetricians and Gynecologists - Medical Specialty Society

Source(s) of Funding

American College of Obstetricians and Gynecologists (ACOG)

Guideline Committee

American College of Obstetricians and Gynecologists (ACOG) Committee on Practice Bulletins-Obstetrics

Composition of Group That Authored the Guideline

Not stated

Financial Disclosures/Conflicts of Interest

Not stated

Guideline Status

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Guideline Availability

Electronic copies: None available

Print copies: Available for purchase from the American College of Obstetricians and Gynecologists (ACOG) Distribution Center, PO Box 933104, Atlanta, GA 31193-3104; telephone, 800-762-2264, ext. 192; e-mail: sales@acog.org. The ACOG Bookstore is available online at the

ACOG Web site .

Availability of Companion Documents

None available

Patient Resources

None available

NGC Status

This NGC summary was completed by ECRI Institute on October 8, 2007. The information was verified by the guideline developer on December 3, 2007. The currency of the guideline was reaffirmed by the developer in 2011 and this summary was updated by ECRI Institute on November 15, 2012. This summary was updated by ECRI Institute on June 1, 2016 following the U.S. Food and Drug Administration advisory on Opioid pain medicines.

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